ISOLATED SCLEROTIC INVOLVEMENT OF THE MITRAL VALVE.

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The frequency of occurrence of mitral-valve deformity and disease independent of and in the absence of other cardiac-valve implication, is one that has received attention only, as a rule, from the standpoint of vegetative endocarditis. It has been described customarily as rheumatic in origin, and at least in its acute forms has been regarded as a part of true bacterial rheumatic disease.

Sclerotic thickening and deformity, causing insufficiency or stenosis, and oftentimes both anatomic effects, have received scant attention at the hands of students of the heart, and perhaps less than has been merited by their actual frequency and interest. Both in the wards of the Philadelphia Hospital and in the autopsy opportunities of private practice I have been impressed by the sclerotic nature of occasional mitral lesions, which I had been led by the history of the case during life and its course while under my observation, to regard as subacute recrudescences of a bacterial endocarditis of a vegetative type. Usually the physical signs have indicated mitral insufficiency or mitral stenosis, or both. Less frequently the indications have been those of purely aortic disease. Irrespective of the location my attention has been attracted by the sclerotic nature of the process and the necessity of inquiring whether I had erred in my understanding of its etiology. I confess that with each new instance, in spite of the association of tonsillitis, joint symptoms, or even pericardial inflammation, I have been forced to look away from acute rheumatism as a satisfactory explanation of the lesion, and to search for a more likely immediate cause.

In this brief study I will refer only to the sclerotic changes of the mitral valve that occur in the absence of apparent lesions of the other cardiac valves. In a later communication I hope to point out the significance of similar processes limited to the aortic valve.

The Anatomical Lesion. I have already referred to the fact that I have at autopsies studied examples of sclerotic insufficiency, also of the frequent combination of sclerotic stenosis with insufficiency of the mitral valve. I have never seen such a stenosis in the absence of an accompanying anatomical insufficiency, though not always of such a character as to furnish the classical auscultatory signs. I have observed a fibrosclerotic insufficiency in the absence of a deformity amounting to an anatomical stenosis, though far less regularly did the patient during life show either the physical signs or the symptoms of valvular obstructive disease. The photographs are illustrative of the isolated sclerotic involvement of the mitral vavles.

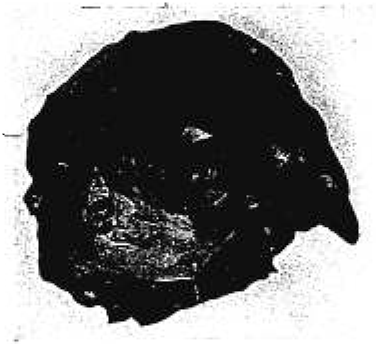

FIG. 1.—High-grade sclerotic involvement of the mitral valve resulting in stenosis and insufficiency.

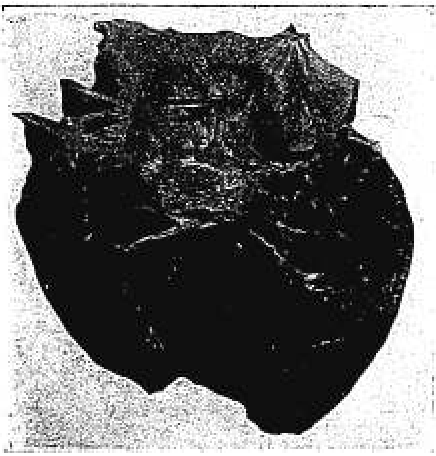

FIG. 2.—Sclerosis and atheroma of the left auricular wall and of the mitral valve.

Microscopic study of the diseased valve leaflets and cusps has shown in the early cases the typical changes of a fibrosclerosis. In more advanced cases atheromatous degeneration is often superimposed, and in a large percentage, lime salts have been deposited, and resulted in a greater or less degree of calcareous change. There has been an entire absence of bacterial findings, and, as far as staining methods are concerned, there has been a complete failure to demonstrate the presence or the causal influence of bacterial forms. I shall later refer to the possibility of the demonstrating spirochetæ in such tissue. The success of a number of laboratory workers in staining the Spirochæta pallida in sections of the aorta that have been in preservative fluids for a number of years leads us to believe that we will yet be able to determine a causal organism in at least those instances that depend upon the Spirochæta pallida for an etiologic factor. All grades of fibrosis of the mitral valve have been observed from the slightest possible thickening to a gross deformity such as is seen in Fig. 2.

THE PHYSICAL SIGNS AND CLINICAL COURSE OF THE DISEASE. I know no clinical method of distinguishing the sclerotic type of valvular disease from the vegetative except in the probable exclusion of syphilis from the etiology by the Wassermann test. In many of the cases that have come under my observation, syphilis has either been admitted by the patient, or there has been sufficient reason to believe that such an admission would be in order. I believe that many instances of hereditary lues are evidenced in childhood by fibrous arteries and by sclerotic involvement of the myocardium and of the valves—the mitral, the aortic, seldom both in the same case prior to late adult years. I reported several years ago a study of a child of four years dying of a rupture of an aortic aneurysm. The aortic valve was so completely closed by sclerotic thickening and deformity as to cause wonder that enough blood could have passed its ring to maintain life. The child's arteries were of the pipe-stem order. The mother was apparently healthy, but the father had a slight degree of saddle-nose, and developed epileptiform convulsions at thirty-one years. He was in all likelihood a case of latent hereditary syphilis or a none too frank instance of the acquired type.

In a few cases of this nature I have neither been able to obtain an admission from the patient that luetic infection has taken place, nor have I had any reason to think the statement of his freedom from infection was incorrect. I have seen cases that clinically resembled tonsillitis and rheumatism, with endocarditic involvement, which later at the autopsy showed an isolated sclerosis of the mitral valve. I remember one little girl that gave only a history of recurrent tonsillar swelling, yet whose arteries and mitral valve were as rigid as those of an old person.

To the question, What is the exact nature of such a case, I reply, I do not yet know. Only a searching study of sections of the arteries and the valve leaflets of such cases will exclude the spirochete, and only the Wassermann test, employed as a routine measure in cardiac disease, will assist in the diagnosis *in vivo.*

FIG. 3.—Sclerosis of the mitral valve. Great hypertrophy of the left ventricular wall.

These sclerotic changes occur in the absence of the classical signs of acquired syphilis. Syphilis not infrequently causes a symptom-complex indistinguishable (except by laboratory means) from that of both acute inflammatory rheumatism and of tonsillitis, and it can and does leave in some instances permanent traces in the form of thickening and deformed mitral leaflets. Whether it is the usual cause of these isolated scleroses is another matter. I have also noted two instances of sclerotic involvement of the mitral valve in sisters, members of a family in which a large number of the recent ancestry had been tuberculous. One parent, the mother, had died in early life, probably though not certainly of tuberculosis. The father died of a systemic nervous disease that may easily have been luetic in origin. The children of this parentage, five in number, have all shown evidence of cardiac valvular disease, which in at least two instances was limited to the mitral valve. Thus we have the possibility of a tuberculous influence and the certainty that syphilis may and sometimes does produce such a picture alone.

Whether other infections may constitute or independently cause isolated valvular sclerosis can only be determined by the studies of many observers of a large series of cases. Dr. James M. Anders informs me that he has studied two interesting cases of this type of mitral involvement in which the causative factor remained unknown. Whether or not acute infectious rheumatism can and ever does cause sclerosis of the valvular tissues is also a point to be considered. As a rule, fibrosis does not form even a prominent feature of the damage consequent upon this disease. Vegetative endocarditis is so generally a rheumatic sequela that this type of valvular disease may almost be regarded as rheumatism of the endocardium. We may therefore pass it over as an unimportant factor in the etiology of the condition under consideration. The text-books have seemingly overlooked mitral sclerosis and implication of the bicuspid valve.

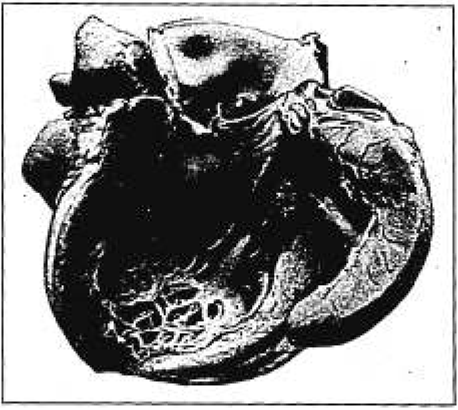

FIG. 4.—Fibrosis of the mitral valve. Marked dilatation of the left ventricle and relative mitral insufficiency. Great hypertrophy of the papillary muscles.

POSSIBILITY OF ANTEMORTEM DIAGNOSIS. From my experience in the cases studied by me I am inclined to believe that all patients, irrespective of age, with sclerosis of the mitral valve, whether associated with other valvular involvements or isolated, will be found also to present sclerotic arteries. The child of four years already mentioned as dying from a ruptured aortic aneurysm had brachial and radial arteries that would have done credit to an intemperate old man. This early arteriofibrosis, together with a premature overaccentuation of the second aortic sound and a tendency to hypertrophy of the left ventricle, will in the presence of the signs of developing mitral disease afford reason to anticipate fibrosis rather than a deposit of lymph and bacteria as the condition of affairs likely to be revealed at the autopsy. A definite history of syphilis or tuberculosis in the patient or in his or her parents will go far toward rendering presumptive evidence certain.

PREVENTIVE MEASURES AND TREATMENT. The only certain preventive measure would be prevention of marriage among the unfit. The question of the right to marry among those who are morally certain to transmit stigmata, if not actual disease, is not a proper subject for a strictly clinical discussion. It has its bearing, however, immediately we begin to consider the prophylaxis of valvular disease.

I doubt whether in the presence of sclerotic arteries in child or adult much can be done in the way of forestalling sclerotic heart change. Both the cardiac muscle and the valve leaflets seem destined to proceed in the course which they have begun before the presence of a toxic factor has been recognized. It is probably too late for us to expect much valvular benefit from antiluetic medication, though in the presence of repeatedly positive Wassermann reactions, thorough courses of arsenic and mercury should be instituted. The studies of Finger and other reliable investigators would seem to indicate that hereditary syphilis is an incurable disease. In the absence of such symptoms as those of lues, we are peculiarly helpless against the insidious march of an irremediable affection. The treatment therefore again reverts in the present very imperfect state of our knowledge of the etiology of these conditions to a new interest in human eugenics, and to precautionary breeding measures that will prove as intelligent in the reproduction of the human species as in the raising of healthy cattle, horses, and hogs.